Fibromatosis: a potential sequela of neuromuscular choristoma

Clinical article

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Object. Neuromuscular choristoma (NMC) is a rare peripheral nerve lesion in which mature skeletal muscle fibers lie within the nerve and its fascicles. Given limited follow-up, its natural history is poorly understood. The occurrence of aggressive fibromatosis in one of the authors’ patients and its occurrence in reported cases suggests an etiological relationship between the 2 lesions. This study attempts to explain the association and its frequency.

Methods. All cases of NMCs seen in consultation or treated at the Mayo Clinic were identified. Demographic and clinical data were reviewed in cases with coexistent aggressive fibromatosis. Pathology and neuroimaging studies were reexamined. In addition, an extensive literature review was performed to explore the association of NMC with aggressive fibromatosis, with special attention given to pathological and imaging characteristics and the development of aggressive fibromatosis.

Results. The authors identified 10 patients with a diagnosis of NMC who were treated at the Mayo Clinic between 1992 and 2010. Four of 5 with adequate follow-up had developed a definite or suspected aggressive fibromatosis. A review of the initial pathological specimens in these cases revealed no evidence of fibromatosis, but all of the lesions exhibited accompanying hypocellular collagenous tissue. On MR images, all cases showed areas of low signal intensity, which significantly differed from muscle, nerve, and NMC components. On available serial MR imaging studies, aggressive fibromatosis seemed to originate in such lower-intensity regions. In the 18 previously reported cases of NMC, 5 patients developed recurrent masses diagnosed as either definite (2 cases) or possible (3 cases) fibromatosis. Review of the published imaging studies in these cases suggests the presence of lower intensity areas similar to those observed in the 10 patients treated at the Mayo Clinic.

Conclusions. This study confirms that the development of aggressive fibromatosis in patients with NMC has been underreported. A direct relationship between the NMC and the development of aggressive fibromatosis is suggested by pathological and neuroimaging evidence. (DOI: 10.3171/2011.6.JNS102171)

KEY WORDS • desmoid tumor • neuromuscular hamartoma • tumor • peripheral nerve

Neuromuscular choristoma is a lesion composed of mature muscle fibers and peripheral nerves in which myocytes are intimately associated with the nerve fibers and are, at least in part, endoneurial. The result is a grossly expanded nerve with multifascicular involvement. Since the first description of this lesion in 1895, approximately 40 cases of NMC are said to have been described. However, some of these cases, especially the ones affecting small intracranial nerves, may represent a different entity. More bona fide examples of NMC generally affect larger nerves such as the sciatic nerve or the brachial plexus. Patients with these lesions typically present in early childhood with a mass and/or neurological symptoms. Although these lesions are described as nonneoplastic, knowledge of their natural history is limited because of their rarity and the short follow-up.

Recently, our 1 previously reported patient29 presented with aggressive fibromatosis (desmoid tumor) fully 8 years after his original diagnosis of NMC. The occurrence of this complication in this patient and in 2 other previously reported cases3,33 raises questions about the pathogenesis of NMC and its etiological relationship with aggressive fibromatosis. In an attempt to explain the association, we reviewed additional cases either evaluated or reviewed at our institution and those reported in
Cases Reported in the Literature

We searched MEDLINE databases using the terms “neuromuscular hamartoma,” “neuromuscular choristoma,” “benign triton tumor,” and “nerve rhabdomyoma,” and cross-referenced all relevant articles to retrieve the greatest possible number of NMC cases. All pathological descriptions and histological samples were reviewed by a neuropathologist (B.W.S.). Cases were included only if sufficient pathological descriptions and/or histological images clearly supported the diagnosis of NMC. The descriptive and/or imaging evidence of largely mature-appearing muscle fibers and peripheral nerve fibers in which myocytes were intimately associated with nerve fascicles and were at least in part endoneurial was required for inclusion of a case in the analysis. In all included cases, patient demographic characteristics, presenting symptoms, imaging data, surgical treatment, and available follow-up information were reviewed. All provided MR imaging images were examined for the presence of areas of signal intensity different from the signal intensity of muscle and/or nerve by the same musculoskeletal radiologist (K.K.A.).

Results

Cases Encountered in our Institution

We identified 10 patients in whom a diagnosis of NMC was established at our institution between 1992 and 2010, 4 of whom had definite or suspected aggressive fibromatosis. One of these 4 cases was previously reported and was revisited given the development of aggressive fibromatosis at recent follow-up. An additional case reviewed at our institution with the diagnosis of both NMC and aggressive fibromatosis is included in the literature review section since it was reported previously.

Of the 6 patients with NMC who did not develop aggressive fibromatosis, only 1 patient had adequate follow-up: a 12-year-old girl, who underwent a subtotal resection of a brachial plexus mass, with no evidence of progression of the residual mass on serial MR imaging studies performed over a 10-year period. One other patient had been treated at our institution; the NMC was of the buccal area, but only 1-year follow-up was available. The other 4 patients were treated at outside institutions; only the pathological specimens (NMC of the forehead, neck, pharyngeal, and unknown site) were reviewed at our institution, and no clinical follow-ups were available.

The 4 patients with NMC and aggressive fibromatosis (Table 1) included 2 males and 2 females presenting between the ages of 5 and 42 years, with the lesions involving the sciatic nerve (3 cases) and brachial plexus (1 case). All presented with a neuropathy or plexopathy confirmed by electrodiagnostic studies, and 3 had secondary musculoskeletal changes (foot deformity [2 cases] and/or extremity asymmetry [2 cases]). The 2 adult patients also presented with progressive pain. The MR imaging studies showed fusiform enlargement of the involved nerve(s). The diagnosis of NMC was made on the basis of either open biopsy (3 cases) or CT-guided biopsy (1 case). Between 4 months and 8 years postoperatively, all patients presented with a new enlarging mass in the vicinity of the NMC and the biopsy site. Biopsy revealed aggressive fibromatosis in 3 of the 4 patients; in the other patient, although imaging was consistent with aggressive fibromatosis, the limited biopsy failed to confirm the diagnosis. Management varied and consisted of observation and resection, chemotherapy followed by resection and/ or radiotherapy at recurrence.
### Table 1: Summary of cases of NMC with definite or suspected aggressive fibromatosis*

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Age (yrs), Sex</th>
<th>Location &amp; Extent</th>
<th>Symptoms &amp; Signs at Presentation</th>
<th>Initial Op</th>
<th>Aggressive Fibromatosis or Recurrence</th>
<th>FU Onset Location Treatment</th>
<th>FU</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tiffee &amp; Barnes, 1998</td>
<td>1.6, F</td>
<td>Lt face &amp; orbit</td>
<td>mass</td>
<td>complete resection</td>
<td>recur: 1 mo postop</td>
<td>Lt face</td>
<td>observation</td>
</tr>
<tr>
<td>Chen, 1984</td>
<td>0, M</td>
<td>Rt median nerve at axilla</td>
<td>mass</td>
<td>complete resection</td>
<td>recur: soon after op</td>
<td>Rt axilla</td>
<td>observation</td>
</tr>
<tr>
<td>Boman et al., 1991</td>
<td>4, F</td>
<td>Lt sciatic nerve at knee area</td>
<td>spasticity &amp; equinism</td>
<td>muscle Bx</td>
<td>1 yr</td>
<td>Lt popliteal area</td>
<td>resection</td>
</tr>
<tr>
<td>Bonneau &amp; Brochu, 1983</td>
<td>5, F</td>
<td>Rt axilla</td>
<td>mass</td>
<td>resection</td>
<td>initial op</td>
<td>Rt axilla</td>
<td>resection</td>
</tr>
<tr>
<td>Mitchell et al., 1995</td>
<td>0, F</td>
<td>Rt axilla &amp; chest wall</td>
<td>mass</td>
<td>partial resection</td>
<td>5 mos postop</td>
<td>Rt axilla &amp; chest</td>
<td>complete resection</td>
</tr>
<tr>
<td>Awasthi et al., 1991</td>
<td>0.6, F</td>
<td>Lt brachial plexus</td>
<td>mass</td>
<td>partial resection</td>
<td>recur: 35 yrs</td>
<td>Lt brachial plexus</td>
<td>resection</td>
</tr>
<tr>
<td>Present report</td>
<td></td>
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<tr>
<td>Case 1</td>
<td>18, M</td>
<td>Rt sciatic nerve from pelvis to lower thigh</td>
<td>neuro: pain, hypesthesia, &amp; weakness in rt sciatic distrib, msk: shorter LE &amp; smaller foot w/ pes cavus &amp; hammer-toes</td>
<td>open fascicular Bx (upper thigh)</td>
<td>8 yrs post-Bx</td>
<td>Rt mid to proximal thigh mass</td>
<td>CT-guided Bx; total resection w/ preservation of sciatic nerve; radiotherapy 15 mos after recur</td>
</tr>
<tr>
<td>Case 2</td>
<td>42, M</td>
<td>Rt sciatic nerve from pelvis to buttock</td>
<td>neuro: deterioration of function &amp; pain, msk: Lt congenital hip dysplasia</td>
<td>CT-guided Bx</td>
<td>3 yrs post-Bx</td>
<td>Lt buttock close to sciatic notch</td>
<td>CT-guided Bx; observation of mass &amp; hip replacement (dysplasia)</td>
</tr>
<tr>
<td>Case 3</td>
<td>5, F</td>
<td>Rt sciatic nerve in mid-thigh area</td>
<td>neuro: weakness in rt sciatic distrib (incl foot drop), msk: cavovarus foot</td>
<td>open fascicular Bx (midhigh)</td>
<td>4 mos post-Bx</td>
<td>Rt mid thigh area (region of previous Bx)</td>
<td>open Bx; 18 mos of chemo; radical resection (mass &amp; sciatic nerve)</td>
</tr>
<tr>
<td>Case 4</td>
<td>6, F</td>
<td>Rt brachial plexus; C4–8 roots to divisions</td>
<td>neuro: weakness in rt UE, msk: smaller UE</td>
<td>open Bx (supraclavicular)</td>
<td></td>
<td></td>
<td>enlarging mass on serial MRI over 16 mos discrete from brachial plexus NMC, which has remained stable</td>
</tr>
</tbody>
</table>

* Bx = biopsy; chemo = chemotherapy; deter = deterioration; distrib = distribution; FU = follow-up; incl = including; LE = lower extremity; msk = musculoskeletal; neurol = neurological; recur = recurrence; UE = upper extremity.
**Retrospective Pathology Review.** In all patients, the initial pathology was that of NMC without evidence of fibromatosis (Fig. 1). However, all lesions were associated with surrounding densely collagenous areas of low cellularity. These were not distributed homogeneously within the specimens. At recurrence, in 3 of the 4 lesions (Cases 1, 2, and 3), the histology of the mass was that of aggressive fibromatosis (Fig. 1). In 2 cases (Cases 1 and 3), the aggressive fibromatosis was histologically in close association with or spatially related to the NMC. This relationship was not evident on the specimen obtained in the limited CT-guided biopsy performed in Case 2. In the last instance (Case 4), the biopsy revealed only hypocellular collagenous tissue, perhaps not representative of the imaging abnormality.

**Retrospective Imaging Review.** In 3 of our 4 patients (Cases 1, 2, and 4) the initial MR imaging study showed areas with signal intensities different from and significantly lower than that of muscle, nerve, and other portions of the NMC. These areas were located within and/or at the periphery of the NMC (Figs. 2–4). Qualitative differences in signal intensity were confirmed quantitatively by calculating ratios of relative signal comparing the area of interest with muscle, nerve, and the rest of the NMC. On serial MR imaging studies available for Cases 1, 2, and 4, the areas with a different signal intensity had persisted and the difference was again confirmed quantitatively with relative signal intensity ratios. When all examinations were considered, the area with a lower signal intensity was significantly different from muscle, nerve, and the rest of the NMC (p < 0.003). In 1 case (Case 3), the initial imaging study was unavailable. However, in limited images available from a subsequent MR imaging study, similar signal intensities different from those of muscle, nerve, and other areas of the NMC were seen at the time of aggressive fibromatosis development (Fig. 5). Based on available serial MR imaging studies in Cases 1, 2, and 4, aggressive fibromatosis seems to form in the vicinity of, or even originate within, these persistent regions of lower intensity.

**Cases Reported in the Literature**

With the search criteria, 50 articles referring to 66 cases of NMC were found. After review, 48 cases were excluded for various reasons: 1) the diagnosis of NMC could not be clearly confirmed because of the lack of pathological information,9–11,13,24,25,32,35,45 2) most cases involving cranial nerves were either difficult to assess because of limited information or seemed to represent a different pathology consisting of muscular and neural elements with a different relationship and often combined with the presence of lipomatous cells;6,14,16–18,23,26,37,39,40,43,46,49,51,53,54 and 3) the histological appearance of the lesion (relationship between muscular and neural elements) differed from classic NMC15,19,34,38,41,48,52 or the lesion was associated with a spinal lipoma.7,47 The reported case by Maher et al.29 is included in the patients encountered at our institution and excluded from the literature analysis. A total of 15 articles were included in this analysis reporting 18 cases.1–5,8,12,20–22,27,30,31,36,42
These patients with NMC presented mostly in early childhood (before the age of 2 in 10 of 18 cases), with only 4 patients presenting in adulthood; the mean age of presentation was 9.75 years (range birth to 68 years). Both sexes were affected, with 11 cases involving female patients and 7 involving male patients being reported. Major peripheral nerves were most frequently involved (8 brachial plexus nerves, 6 sciatic nerves), but lesions were also reported to occur in intercostal nerves, the vaginal wall, the face, and arytenoid submucosa. Although brachial plexus lesions could involve multiple nerves, all lesions were solitary except for 1 case in which there were 2 dis-
distinct masses along the same nerve. Nine of the patients presented initially only with a mass. The case involving 2 masses along the same nerve was identified at autopsy. In all the other cases, the patients had symptoms related to mass effect—stridor (in 1 patient), pain (in 2 patients), sensory and/or motor deficits (in 3 patients), and/or secondary musculoskeletal deformity (in 2 patients). In all cases except the one found at autopsy, the patients underwent surgery. The initial surgery was biopsy in 4 cases, partial resection in 7, and complete resection in 6.

Recurrences and Occurrence of Aggressive Fibromatosis. In the literature, patients with NMC were followed up for a mean of 2.9 years (median 2.75 years), with the duration of follow-up ranging from the immediate postoperative period to 9 years postoperatively. Of the 18 patients, 6 developed a recurrent mass. Only 1 patient with a recurrent mass did not have evidence of aggressive fibromatosis: the recurrent or possibly residual tumor was seen on imaging 1 month postoperatively and remained stable over the following 2.5 years. The other 5 cases with a recurrent mass either clearly (2 cases) or probably (3 cases) developed aggressive fibromatosis. In 1 case, a recurrence noted soon postoperatively remained stable for 4 years. The area was then reexplored, but because the mass was involving the median nerve, only a biopsy was performed. The biopsy showed dense fibrous tissue, which potentially could have represented aggressive fibromatosis. The mass remained stable for the subsequent 2 years of follow-up. In the 3 other patients, the mass recurred after biopsy (1 year), resection (1 year), or partial resection (1 month). One or more additional resections were performed in each case, but the masses kept recurring, ultimately requiring amputation. In all 3 cases, pathology specimens revealed NMC and aggressive fibromatosis. In 2 of the cases, aggressive fibromatosis was initially seen (although not diagnosed per

![Fig. 3. Case 2. Magnetic resonance images obtained preoperatively (A–C) and 5 years after CT-guided biopsy (D–F). A: Axial T2-weighted image with fat suppression at the level of the acetabulum shows a mass (arrow) with heterogeneous but overall low signal. An area of lower signal consistent with fibromatosis (arrowhead) is seen at the periphery of the mass encasing the sciatic nerve. B: Axial T1-weighted image at the same level shows the mass with low signal extending into the sciatic notch. Note atrophy in the gluteus maximus muscle (plus signs). C: Axial Gd-enhanced T1-weighted image with fat suppression shows enhancement of the mass (arrows) with some focal, nonenhancing low signal areas. The enhancement within the gluteus maximus muscle is related to subacute denervation change. D: Axial T1-weighted image at the level of the ischial tuberosity shows enlargement the heterogeneously low signal mass, which is now causing very significant mass effect on the adjacent gluteus maximus muscle (plus signs). E: Axial T1-weighted image obtained slightly superior to D showing the increase in size of the mass as well as further intrapelvic extension. Again noted are sheetlike areas of low signal representing fibrous tissue (arrowheads). Total hip arthroplasty causes artifact on this image. F: Coronal T1-weighted image shows the full extent of the mass through the sciatic notch as well as the prominent muscular atrophy.](image-url)
Fibromatosis following neuromuscular choristoma

In these 2 cases, NMC was only diagnosed at amputation or retrospectively. In the other case, NMC was initially diagnosed, and aggressive fibromatosis was noted at recurrence. One additional patient could also be considered to have had a recurrent mass and possible aggressive fibromatosis: a previous partial resection of a mass in the same area, likely of the same pathological type, was done 35 years earlier when the patient was 7 months old. The initial pathological examination revealed dense fibrous tissue interdigitating with skeletal muscle; 35 years later, histological examination revealed neuromuscular elements admixed with dense fibrous tissue.

Pathology and Imaging Review. In all included cases, the pathological findings were consistent with NMC. The reports described nerve fibers and mature skeletal muscle. However, in 11 of the 18 cases the report also mentioned a component of increased, dense fibrous tissue, bands thereof, or a collagenous matrix. Magnetic resonance images provided in 6 of the 18 cases of NMC all exhibited areas of very low signal intensity different from the signal intensity of muscle and/or nerve. These regions were compatible with the usual MR imaging appearance of fibrous tissue.

Discussion

Despite various theories regarding the pathogenesis of NMC, its true nature and the mechanisms underlying its development remain to be established. Little is known of its natural history and evolution over time. Our report emphasizes the occurrence of aggressive fibromatosis in NMC. A potential etiological relationship has only been briefly mentioned. Its basis remains elusive. Questions remaining include: 1) Is the occurrence of aggressive fibromatosis in patients with NMC a coincidence? 2) Is the development of aggressive fibromatosis in these patients related to surgery? 3) Can the development of aggressive fibromatosis be related to other factors? and 4) Does a direct relationship exist between NMC and the development of aggressive fibromatosis?

Is the Occurrence of Aggressive Fibromatosis in Patients With NMC a Coincidence?

The occurrence of aggressive fibromatosis in patients with NMC seems too high to be a coincidence. If the cases encountered at our institution (10 cases) and the cases reported in the literature (18 cases) are combined, at least 5 patients had aggressive fibromatosis (Cases 1–3 and 2 cases reported in the literature), and 4 others likely did (Case 4 and 3 cases reported in the literature). This means that aggressive fibromatosis occurred in 18% to 32% (5/28 to 9/28) of patients. In the context of a short follow-up for the cases reported in the literature (mean of 2.9 years) and the delayed development of fibromatosis in
some patients (up to 8 years in Case 1), it is possible that
the occurrence of fibromatosis in these patients may be
even higher. Within the literature, 48 cases have been ex-
cluded, largely because these cases were not clearly NMC
and may have represented other pathological processes.
However, even if these cases were all considered to be
NMC, and all did not develop aggressive fibromatosis,
aggressive fibromatosis would have still occurred in 7%
to 12% (5/76 to 9/76) of patients. Since both lesions are
rare, it is fair to say that the development of aggressive
fibromatosis in NMC patients is more than a coincidence.

Is the Development of Aggressive Fibromatosis in These
Patients Related to Surgery?

The etiology of aggressive fibromatosis is poorly un-
derstood, but the development is thought to be influenced
by physical factors such as trauma and surgery,28,44 espe-
cially in patients with familial adenomatous polyposis.44
In some of our patients, surgery seems to be an initiating
event. Clearly, in 3 of our cases (Cases 1–3) and in 2 of the
cases described in the literature,4,33 surgery preceded and
perhaps promoted the development of the fibromatosis.
However, fibromatosis was present in one of the cases at
the time of the initial surgery. Therefore, surgery may
frequently be an initiating factor, but other mechanisms
are also likely involved.

Can the Development of Aggressive Fibromatosis in These
Patients be Related to Other Factors?

Hormonal factors have been reported to influence
the pathogenesis of fibromatosis. For example, abdomi-
nal fibromatosis mainly occurs in female patients during or
after pregnancy.44 In patients with NMC, fibromatosis de-
veloped in females (6 cases) as well as in males (3 cases),
and mainly in childhood (6 cases). No endocrine abnor-
mality has been reported, but it is difficult to eliminate
possible hormonal influences. In the literature, fibromato-
sis has also been associated with both Gardner syndrome
and familial adenomatous polyposis, thus suggesting a
genetic predisposition for the development of such tu-
mors. In patients with NMC, a genetic susceptibility to
the development of aggressive fibromatosis is also pos-
sible. However, the fact that all aggressive fibromatosis
develops in the area of the NMC suggests that a genetic
predisposition is unlikely to be the single or dominant
factor leading to their development.

Does a Direct Relationship Exist Between the NMC and
Development of Aggressive Fibromatosis?

In patients with NMC, we believe that there is a di-
rect relationship between the occurrence of aggressive fi-
bronatosis and NMC, perhaps in addition to other factors
discussed above. In support of this notion is the observa-
tion that all patients who developed aggressive fibroma-
tosis have done so at the site of their NMC. Moreover,
histological examination of the pathological specimens
showed the aggressive fibromatosis to be histologically in
close spatial association to the NMC elements in 2 of our
cases and in 2 previously reported cases.4,5

Our pathological review has shown that NMC is,
even initially, somewhat heterogeneous in makeup. In
addition to the variable combination of nerve and ske-
enetal muscle fibers, most lesions, both at our institution
and in the literature, possess variable amounts of fibrous
and/or collagenous tissue. The heterogeneity of NMC,
even at initial presentation, is confirmed on imaging by
the presence or emergence of signal intensities differing
from those of muscle and/or nerve. Both our cases and
the available images of published cases show this signal
heterogeneity, possibly reflecting the presence of fibrous
tissue, even at an early stage. In our cases, serial imaging
suggests that the aggressive fibromatosis develops in rela-
tion to these areas of signaling. It appears that fibroma-
tosis develops within or at the periphery of the NMC at an
early stage and the process of fibrous tissue proliferation
is activated by a surgical procedure. This is not to say that
simple chronic fibrosis foreshadows the development of
fibromatosis. Lastly, the absence of fibromatosis at often
limited initial biopsies does not preclude its presence. In
that quite frequently only a fascicular biopsy is taken, a
fibromatosis component may well have been unsampled.

Various theories regarding the etiology of NMC
could be consistent with a direct relationship between
NMC and the development of aggressive fibromatosis.
Orlandi26 suggested that NMC results from the entrap-
ment of muscle (or mesenchymal cells) into the substance
of developing nerves; other mesenchymal cells, such as fi-
broblasts, could also be incorporated. Masson31 suggested
that neuroectoderm was capable of mesenchymal differ-
etiation, specifically skeletal muscle. Even arachnoidal
cells comprising the leptomeninges are, in part, neuroec-
todermal in derivation.50 This mechanism could explain
the development of NMC-associated fibromatosis in that
differentiation of primitive cells toward fibroblasts may
also be possible. Finally, it has been suggested that
the development of fibrosis may be related to degeneration
of neural components and thereafter degeneration of the
skeletal muscle component due to loss of trophic stimu-
li.20 This process could lead, depending upon the effect of
other factors, to spontaneous regression of NMC or to the
development of aggressive fibromatosis. Just which mech-
anism is responsible for the development of NMC and its
association with aggressive fibromatosis is currently un-
known, but evidence suggests that various mechanisms
likely facilitate the process.

Conclusions

The present study confirms that aggressive fibro-
matosis in patients with NMC has been underreported.
Pathological and imaging evidence suggests that a direct
relationship exists between NMC and the development of
aggressive fibromatosis. Further careful follow-up study
may help determine the best preventive and therapeutic
approaches to aggressive fibromatosis.

Addendum

Since the submission of this manuscript, the senior author has
treated yet another patient with NMC unassociated with aggressive
fibromatosis (desmoid tumor). This 14-year-old boy had a long his-
tory of progressive sciatic neuropathy with leg-length discrepancy,
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a cavus foot, and an enlarged sciatic nerve, features reminiscent of a previous patient described in an earlier publication.29 The diagnosis of NMC, suspected based on clinical history and physical examination, was supported by imaging findings. The MR imaging abnormality extended within the sciatic nerve from the proximal thigh through the sciatic notch to involve the S-1 nerve, and the lesion had both adipose and soft tissue elements. Collectively the features suggested NMC, rather than lipomatosis of nerve (fibrolipomatous hamartoma), in that 1) undergrowth of the affected limb, rather than the overgrowth often seen in patients with lipomatosis of nerve, as well as 2) relative paucity of intraskeletal adipose tissue on MR imaging, were noted. Intraoperative stimulation of the tibial and peroneal divisions of the sciatic nerve and of a hamstring branch resulted in contraction of the nerve(s) (Video 1), thus confirming the diagnosis of NMC in which myocytes are present within nerve. A fascicular biopsy confirmed the diagnosis of NMC unassociated with aggressive fibromatosis. The specimen, a hamstring branch, consisted of a grossly brown nerve—the color being a reflection of the muscle content. Histologically, the mature muscle fibers were intimately admixed with nerve fibers. Long-term follow-up will be necessary in order to detect the possible development of fibromatosis, which may occur many years after diagnosis of an NMC. Based on our experience with this condition, we anticipate that the diagnosis of NMC can be suggested by careful interpretation of MR images and correlation with the clinical information. Establishing the diagnosis with a high degree of confidence using imaging criteria may eliminate the risk of surgery inducing a fibromatosis.

Click here to view the video with Windows Media Player. Click here to view the video with Quicktime.

Disclosure

Dr. Spinner is a consultant for Mayo Medical Ventures. The remaining authors report no other conflicts of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Spinner, Hébert-Blouin. Acquisition of data: all authors. Analysis and interpretation of data: Spinner, Hébert-Blouin, Scheithauer, Amrami. Drafting the article: Spinner, Hébert-Blouin. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Spinner. Statistical analysis: Hébert-Blouin.

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